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# Editorial correspondence

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## Pituitary adenoma in McCune-Albright syndrome: Follow-up information

*To the Editor:*

In the December, 1975, issue of THE JOURNAL OF PEDIATRICS, Dr. Penny, Dr. Frasier, and I reported studies on a 5-5/12-year-old white male with McCune-Albright syndrome, precocious puberty, and gigantism.[1] The findings in our patient were interpreted as evidence for abnormal hypothalamic function, as a cause of the sexual precocity and gigantism. Since the diagnosis at age 3 years, sequential skull roentgenograms for evaluation of the size of the sella turcica remained unchanged. In addition, ophthalmologic and neurologic examinations had shown no change in the patient's status. As recently as July, 1975 (age 7-3/12 years), an ophthalmologic examination revealed normal visual acuity and normal visual fields. In December, 1975 (age 7-8/12 years), however, both optic discs were extremely pale and visual acuity in the right eye was 20/400. In addition, examination of visual fields showed a central scotoma extending 35° temporally in the right eye. The patient was not aware of any visual changes and was asymptomatic. By tomography, the optic foramena were found to be almost completely obliterated by the bony changes in the skull secondary to the McCune-Albright syndrome. It was our opinion that the pale optic nerves were due to compression by the bony changes in the skull. A right frontal craniotomy was performed; the right optic nerve was severely compressed and its course markedly altered by the bony abnormalities. Following this procedure, computerized axial-tomography was unsuccessful because the patient's large head would not fit into the equipment. Therefore, a pneumoencephalogram was performed. A 1.5 cm spherical suprasellar mass which did not appear to involve either the floor of the third ventricle or the hypothalamus was found. Following the diagnosis of the suprasellar mass, a left frontal craniotomy was performed. The mass arose from within the pituitary gland and extended upward through the diaphragmatic sella, exerting mild pressure on the optic chiasma. The mass was biopsied and identified by frozen section to be a pituitary tumor. Excision of the tumor and a hypophysectomy were performed. Subsequent pathologic examination of the mass revealed it to be an eosinophilic adenoma of the pituitary. The left optic nerve was also markedly compressed by the bony changes in the skull and deviated from its normal course. Decompression of the nerve was carried out. Although we initially considered a pituitary adenoma as a possible diagnosis in our patient, previous normal sella roentgenograms, ophthalmologic, neurologic, and visual field examinations did not support this diagnosis. However, the recent clinical changes led to the diagnosis. Recently, it has been proposed that autonomously functioning endocrine glands, such as seen in multiple endocrine adenomatoses syndromes, may explain the endocrinopathies in McCune-Albright syndrome.[2] We cannot say whether our patient's pituitary adenoma was primary and autonomous, or whether it was secondary to prolonged abnormal stimulation by the hypothalamus.[3]

**See related article, p. 73.**

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